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CASE REPORT

Desmoplastic fibroblastoma mimicking tenosynovial giant cell tumor encasing a tendon of the foot

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Abstract

Desmoplastic fibroblastoma is an uncommon, benign fibrous soft tissue tumor that usually occurs in the arms, shoulders, neck, hands, and feet in the fifth to seventh decades of life. In general, it is commonly located in the subcutaneous tissue and skeletal muscle. The authors report an unusual case of a desmoplastic fibroblastoma mimicking tenosynovial giant cell tumor encasing a tendon of the foot in a 72-year-old woman. Ultrasonography revealed an inhomogeneously hypoechoic lobulated soft tissue lesion completely wrapped around the extensor digitorum longus tendon. Color Doppler study revealed increased vascularity in the internal and peripheral portions of the lesion. Magnetic resonance imaging revealed a well-defined, lobulated soft tissue mass encasing the extensor digitorum longus tendon with predominantly isointense signal with some areas of hypointense signal on T1-weighted images, predominantly hyperintense signal with some areas of hypointense signal on T2-weighted images, and inhomogeneous enhancement on fat-suppressed contrast-enhanced T1-weighted images. Surgical excision was performed, and the mass was diagnosed on pathological examination as a desmoplastic fibroblastoma. There has been no previously published radiologic case of a desmoplastic fibroblastoma encasing a tendon of the foot in the literature.

Keywords Desmoplastic fibroblastoma · Collagenous fibroma · Soft tissue tumor · Tenosynovial giant cell tumor · Tendon · Foot

Introduction

Desmoplastic fibroblastoma, also known as collagenous fibroma, is an uncommon, firm, slow-growing, painless, benign fibrous soft tissue tumor [1]. It usually occurs in the fifth to seventh decades of life, with a significant male predominance [2]. It has a wide anatomical distribution; however, the arms, shoulders, neck, hands, and feet are relatively frequent sites [2, 3]. It is usually located in the subcutaneous tissue and skeletal muscle [2]. Rare locations for desmoplastic fibroblastoma include the joint, oral cavity, dermis, lower abdomen, and the parotid and lacrimal glands [4–10]. To the best of our knowledge, there have been no radiologic reports of desmoplastic fibroblastoma mimicking tenosynovial giant cell tumor encasing a tendon of the foot.

The natural evolution of desmoplastic fibroblastoma is characterized by the absence of changes in patterns on magnetic resonance imaging (MRI) despite increases in size, resembling a slow-growing benign tumor rather than an aggressive entity [11]. Unlike tenosynovial giant cell tumor, desmoplastic fibroblastoma has no reported incidence of local recurrence or metastases after surgical excision [12]. Therefore, preoperative radiological diagnosis of desmoplastic fibroblastoma is important to avoid over-treatment and unnecessary extensive procedures [13]. Herein, we report an unusual case of a desmoplastic fibroblastoma of the foot in a 72-year-old woman, which completely encased the extensor digitorum longus tendon.

Case report

A 72-year-old woman presented with a 7-month history of a gradually growing palpable lump on the dorsum of her right

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foot. There was no history of antecedent trauma or local signs of infection. She had been taking oral medication for diabetes mellitus and hyperlipidemia for the past 8 years, and has had hypothyroidism for the past 2 years. There was no other relevant medical history. On physical examination, the lesion was found to be a non-movable, firm, and painless mass, with no tenderness or associated skin changes, detected on the dorsum of the foot. The results of laboratory investigations were within normal limits, except for glycated hemoglobin level (6.9%).

Radiography revealed a relatively well-defined, lobulated, increased soft tissue density on the dorsum of the forefoot at the second metatarsal area. There was no erosion of the underlying metatarsal bones (Fig. 1). Ultrasonography revealed an inhomogeneously hypoechoic lobulated soft tissue lesion along the extensor digitorum longus tendon of the second toe at the level of the metatarsal shaft to head. The lesion was completely wrapped around the extensor digitorum longus tendon, which was located eccentrically to the lateral side within the lesion. Color Doppler study revealed increased vascularity in the internal and peripheral portions of the lesion, which was mildly compressible. There was no communication between the lesion and the joint space (Fig. 2). MRI revealed a well-defined, lobulated soft tissue mass, measuring approximately 3.7 × 2.4 × 2.0 cm in size, along the extensor digitorum longus tendon of the second toe at the level of the metatarsal shaft to the metatarsophalangeal joint. The mass encased the extensor digitorum longus tendon; however, there was no demonstrable abnormality in the tendon. The mass exhibited predominantly isointense signal relative to the adjacent muscle with some areas of hypointense signal on fast-spin echo T1-weighted images, and predominantly hyperintense signal with some areas of hypointense signal on fast-spin echo T2-weighted images. The mass was surrounded by a capsule of hypointense signal. Fat-suppressed contrast-enhanced T1-weighted images demonstrated peripheral and nodular enhancement at the early stage of contrast enhancement and gradually more diffuse inhomogeneous enhancement over time (Fig. 3). There was no evidence of abnormal communication of the lesion into the joint space. The underlying bone had no abnormal findings. On the basis of radiological findings, the initial differential diagnosis of the mass included tenosynovial giant cell tumor.

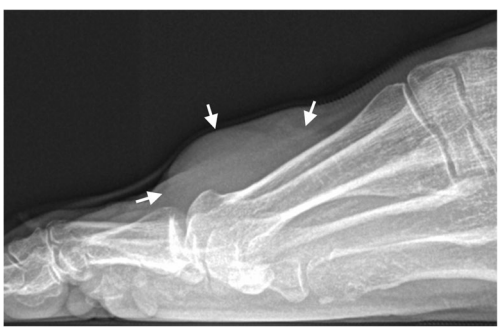

Fig. 1 Radiographs of the foot. Lateral view reveals a relatively well-defined, lobulated, increased soft tissue density (*arrows*) on the dorsum of the forefoot, the second metatarsal area. There is no erosion of the underlying metatarsal bones

Surgical excision of the mass was performed without postoperative complications. During the operation, the lobulated mass was found to envelop the extensor digitorum longus tendon sheath on the dorsum of the foot (Fig. 4). The mass was completely removed by careful dissection, and measured approximately 4 cm in size. The extensor digitorum longus tendon was intact.

The excised mass was submitted for pathological examination as a firm lump measuring 3.5 × 2.0 × 1.5 cm in size. Grossly, it exhibited a well-circumscribed, homogeneously white, lobulated, firm mass with glistening cut surface on sectioning. Microscopically, the mass exhibited well-defined, rounded, smooth contours, and hypocellularity comprising abundant collagenous matrix with low vascularity, stellate-shaped spindle cells with bland nuclei, and no mitotic figures. The mass was ultimately confirmed to be desmoplastic fibroblastoma (Fig. 5).

Discussion

Desmoplastic fibroblastoma is a unique form of benign fibrous soft tissue tumor, which was first described by Evans in 1995 [1]. It has typically been described as a painless, slowly growing mass, often of relatively long duration. It ranges in size from 1 to 20 cm (median 3.0 cm). Histopathologically, the lesion is hypocellular and consists of relatively bland stellate- and spindle-shape cells embedded in a dense collagenous or myxocollagenous stroma. Mitotic figures are absent or rare, and necrosis is absent [2]. Therefore, this tumor is alternatively known as collagenous fibroma because it is clinically and morphologically distinct, as well as completely benign, as described in a previously reported series [14]. The lesions are predominantly subcutaneous; nevertheless, fascial involvement is not rare, and 27% of cases involve skeletal muscle [2]. However, there has been no radiologic report of a desmoplastic fibroblastoma encasing a tendon of the foot, mimicking a tenosynovial giant cell tumor, in the English-language literature.

Radiological findings for desmoplastic fibroblastoma are primarily solid mass without calcification. Rarely, a highly calcified mass or the presence of scattered dystrophic calcification has been reported [15, 16]. To the best of our knowledge, ultrasonography for desmoplastic fibroblastoma has been reported in only one case to date [17]. According to Bonardi et al. [17], imaging revealed a solid mass, isoechoic to muscle with a few inhomogeneous hypoechoic spots, with smooth and lobulated margins. It was localized in the fat, deep under the deltoid muscle and exhibited an expansive rather than an infiltrative behavior, apparently compressing the muscle itself at this point. These

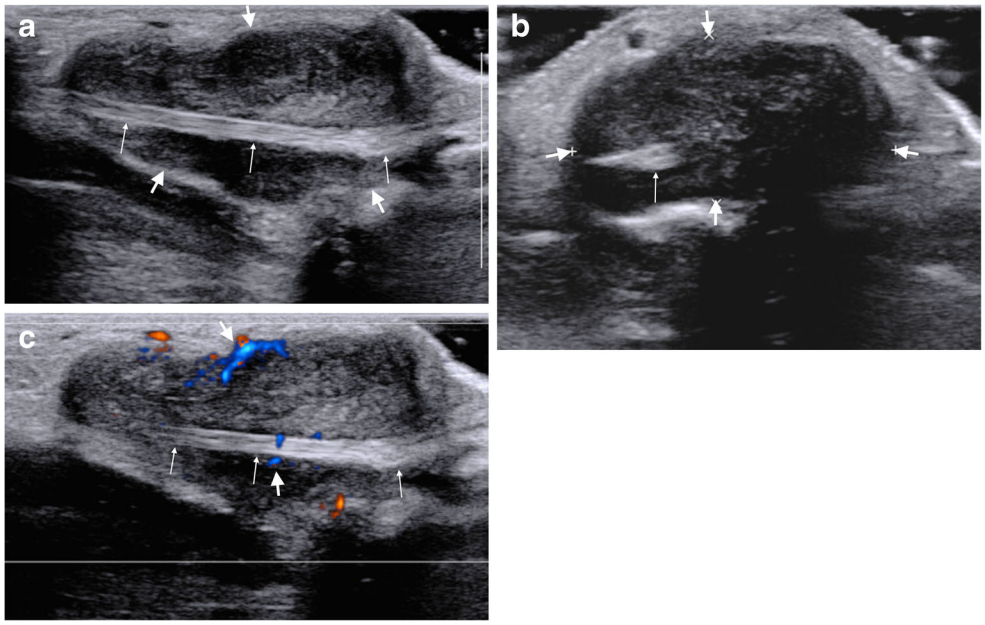

Fig. 2 Ultrasonography of desmoplastic fibroblastoma. Longitudinal view (**a**) reveals an inhomogeneously hypoechoic lobulated soft tissue lesion (*arrows*) along the extensor digitorum longus tendon (*long thin arrows*) of the second toe at the level of the metatarsal shaft to head. On transverse view (**b**), the lesion (*arrows*) is completely wrapped around the extensor digitorum longus tendon (*long thin arrow*). The extensor digitorum longus tendon is located eccentrically to the lateral side within the lesion. Color Doppler study (**c**) reveals increased vascularity (*arrows*) in the internal and peripheral portions of the lesion. (**c**, *long thin arrows*, extensor digitorum longus tendon)

ultrasonographic findings were different from echogenicity in our case, which was lobulated and inhomogeneously hypoechoic relative to the adjacent subcutaneous fat, soft tissue lesion. However, the fact that the masses exhibit an expansive—rather than infiltrative—behavior was similar to our case, in which the tumor was surrounding the tendon but the tendon was intact. During the operation, it was evident that the tumor did not originate from the tendon sheath but was only wrapped around the tendon sheath. Power Doppler evaluation displays a rich diffuse vascularization of the mass [17]. However, our case exhibited increased vascularity in the internal and peripheral portions of the lesion showing distinct hypervascular and hypovascular regions.

MRI findings for desmoplastic fibroblastoma have been described in detail in several reports [8, 11, 13, 18]. Desmoplastic fibroblastomas exhibit inhomogeneously low to intermediate signal intensity on T1-weighted images, inhomogeneously low to slightly high signal intensity on T2-weighted images, and poor or mild or heterogeneous enhancement with areas of no enhancement on contrast-enhanced T1-weighted images [8, 11, 13, 18]. Thus, MRI findings vary among individual tumors because of variable cellularity. Areas exhibiting high signal intensity on T2-weighted images correspond to hypercellular areas within the lesion that consisted of loose collagen fibers. The area of low signal intensity on both T2- and post-contrast T1-weighted images correspond to a hypocellular component with abundant collagen fibers [8, 18].

In our case, preoperative radiological diagnosis was difficult. This is because the mass surrounded the tendon, as observed for tenosynovial giant cell tumors, but had many high signal intensities on T2-weighted images and heterogeneous enhancement on post-contrast T1-weighted images. However, the most likely differential diagnosis was tenosynovial giant cell tumor based on the location of the lesion, signal intensity, and enhancement pattern. Tenosynovial giant cell tumors appear most often as a homogeneous hypoechoic mass adjacent to a tendon on ultrasonography. The adjacent tendon is usually normal in appearance [19]. Although not exclusively, tenosynovial giant cell tumors arising from the tendon sheath uncommonly surround totally the tendon and tend to be more peripheral. However, in general, the sheath of extensor tendons of the foot stops some centimeters proximally to the location of our tumor. On color or power Doppler images, tenosynovial giant cell tumors have substantial flow (abundant flow in the tumor) in most lesions and occasionally minimal flow (sparse flow in the tumor) [20], while our case exhibited increased vascularity in the internal and peripheral portions of the lesion showing distinct hypervascular and hypovascular regions. On MRI, tenosynovial giant cell tumor appears as a focal mass, often adjacent to or surrounding a tendon. It is usually of intermediate or low T1 and T2 signal intensity and is well enhanced with contrast medium administration; however, heterogeneous high T1 and T2 signal intensity can also be apparent in these lesions. If there is sufficient hemosiderin deposition within the mass, it can exhibit

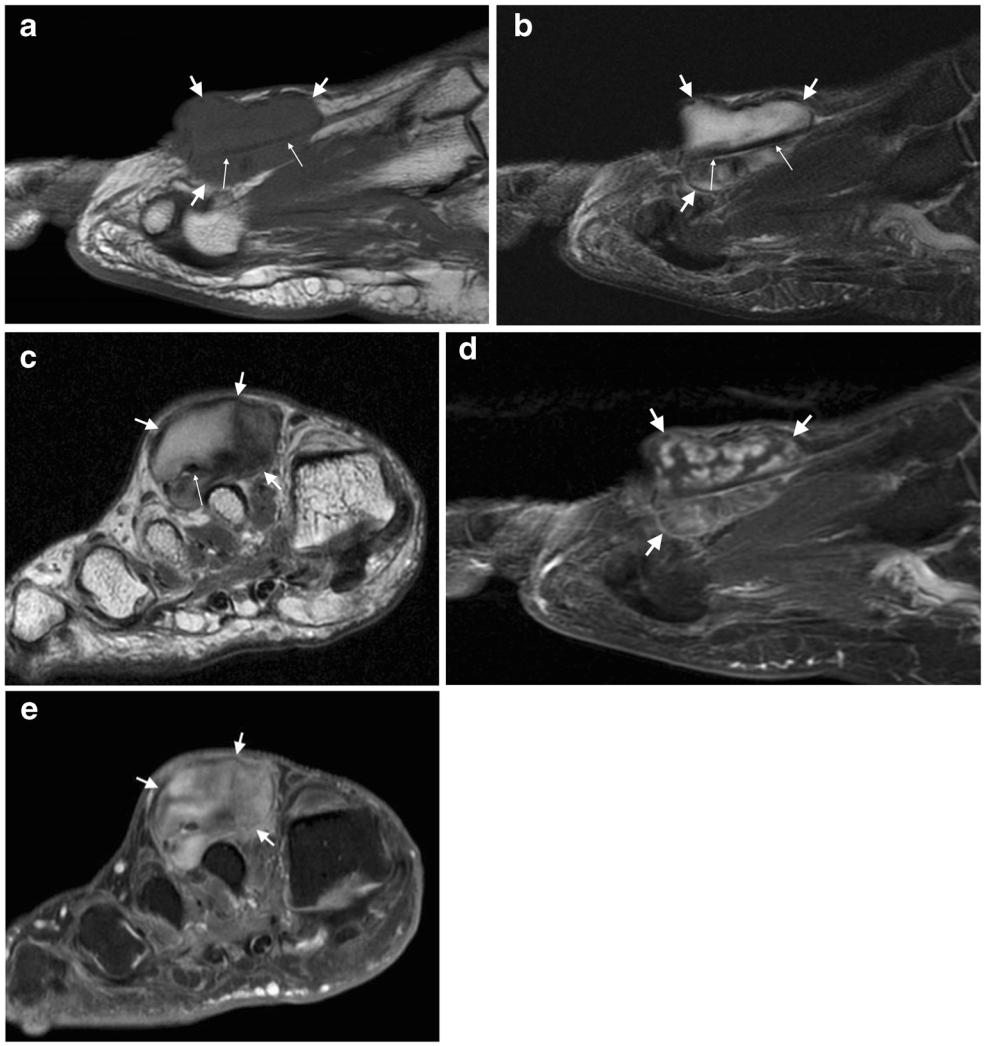

Fig. 3 Magnetic resonance imaging of desmoplastic fibroblastoma. **a-c** A well-defined lobulated mass (*arrows*) encasing the extensor digitorum longus tendon (*long thin arrows*) of the second toe is observed at the level of the metatarsal shaft to the metatarsophalangeal joints. The mass exhibits predominantly isointense signal with some areas of hypointense signal on fast spin echo (FSE) T1-weighted sagittal image (**a**, repetition time [TR]/echo time [TE] = 850/11 ms), and predominantly hyperintense signal with some areas of hypointense signal on FSE fat-suppressed T2-weighted sagittal (**b**, TR/TE = 4310/69 ms) and T2-weighted iterative decomposition of water and fat with echo asymmetry and least-squares estimation (IDEAL) in-phase coronal (**c**, TR/TE = 3556/59 ms) images. The mass was surrounded by a capsule of hypointense signal (*arrows*). **d** and **e** The mass (*arrows*) exhibits peripheral and nodular enhancement at the early stage of contrast enhancement on fat-suppressed contrast-enhanced T1-weighted sagittal image (**d**, TR/TE = 762/16 ms) and gradually more diffuse inhomogeneous enhancement over time on contrast-enhanced T1-weighted IDEAL water-only coronal image (**e**, TR/TE = 770/12 ms)

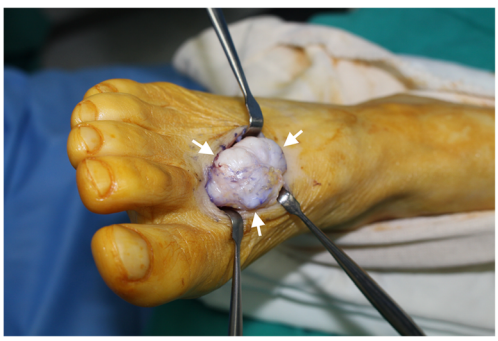

Fig. 4 Surgical excision of the mass. During the operation, the lobulated mass (*arrows*) was found to envelop the extensor digitorum longus tendon sheath on the dorsum of the foot

blooming on gradient-echo MRI [21, 22]. Therefore, although the tumor in this case did not exhibit MRI signal intensity, commonly found in tenosynovial giant cell tumors, we considered tenosynovial giant cell tumor as a first choice in the differential diagnosis because it surrounded the tendon sheath. Other differential diagnoses of the lesion adjacent to tendon sheaths include fibroma of the tendon sheath and inflammatory tenosynovitis such as rheumatoid tenosynovitis.

In conclusion, although a desmoplastic fibroblastomas are usually located in the subcutaneous tissue and skeletal muscle, they may manifest as a mass encasing a tendon of the foot, as in this case; thus, it should be included in the differential diagnosis of a benign mass.

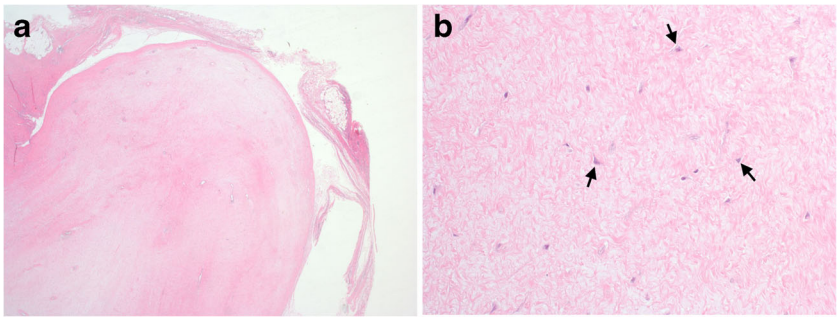

Fig. 5 Histological features of desmoplastic fibroblastoma. **a** The mass exhibits well-defined, rounded, smooth contours (hematoxylin and eosin staining, original magnification ×12.5). **b** The tumor is a hypocellular mass with abundant collagenous matrix and low vascularity. The tumor exhibits stellate-shaped spindle cells (*arrows*) with bland nuclei, and no mitotic figures (hematoxylin and eosin staining, original magnification ×200)

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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